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# Optic Chiasm Glioma, Electrolyte Abnormalities, Nonobstructive Hydrocephalus and Ascites

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A 4-year-old girl with optic chiasm glioma (OCG), nonobstructive hydrocephalus and ventriculoperitoneal shunt is described, in whom marked ascites developed. The ascitic fluid was protein-rich and its amount correlated with cerebrospinal fluid (CSF) protein. The CSF protein level and the amount of ascitic fluid were influenced by chemotherapy. Very unusual hypernatremia, up to 190 mEq/l, with no associated alteration in mental status, was also found. It is suggested that altered absorption ability owing to the high protein content was the cause of both the nonobstructive hydrocephalus and the ascites. The unusual well being with very high sodium concentrations may have resulted from osmoreceptor dysfunction, presumably caused by hypothalamic involvement as well as by the high CSF protein. This combination of findings may point toward specific characteristics of OCG. In an effort to reduce the amount of the ascitic fluid, a further chemotherapeutic trial may be done, before converting the shunt to the vetriculoatrial system. Med. Pediatr. Oncol. 29:33–35,1997. 

**Key words:** CSF ascites; hypodipsic hypernatremia; hypothalamic/chiasmatic glioma; V-P shunt

## INTRODUCTION

The clinical manifestations of optic chiasm glioma (OCG) can result from tumoral involvement of several systems, either by primary penetration or by secondary pressure. Thus, visual, endocrine, cerebrospinal fluid (CSF) obstruction, and ischemic signs and symptoms may occur [1]. When CSF shunting becomes necessary, the clinical picture may also include abdominal problems related to the shunt, for instance, CSF loculations, bowel perforation, fistula formation and catheter tip migration [2].

In the present communication, a girl with unusual complications of OCG is described: marked, disabling, postventriculoperitoneal shunting ascites which improved with chemotherapy, and severe hypernatremia. Similar cases have been previously reported [1,3]. It is suggested that this combination of findings may result from specific characteristics of the OCG.

## CASE REPORT

A 4-year-old girl, a new immigrant to Israel, was referred to our center for treatment of a large OCG. Her family history was unremarkable. Her first two years of life were marked by recurrent episodes of vomiting; later, at the age of 2½ years, visual loss and seizures were noted. After detection, the tumor was partially resected. One year later, because of tumor regrowth and the appearance of hydrocephalus, the patient underwent another resection, and a lumboperitoneal shunt was placed. Later, a ventriculoperitoneal shunt was also installed.

The physical examination at presentation in our center revealed an obese girl with no signs of neurofibromatosis, but with precocious telarche and pubarche. She was blind in the left eye, and visual acuity in the right eye was decreased. Muscle tone was increased and deep tendon reflexes exaggerated. She had marked motor clumsiness.

Neuroimaging studies revealed a large polycystic tumor invading the chiasm, hypothalamus and third ventricle (Fig. 1). The pathology slides were reviewed and confirmed the diagnosis of a low-grade OCG. As her visual fields and visual acuity continued to deteriorate, the tumor was irradiated and chemotherapy with vincristine 1.5 mg/m$^2$ × 10 weeks and VP-16 100 mg/m$^2$ × 5 days was initiated.

One year later, marked weight gain as well as abdominal distention, with evidence of ascitic fluid, were found (Fig. 2). Laboratory evaluation at this stage revealed BUN 8 mg/dl, creatinine 0.6 mg/dl, sodium 142 mEq/l, potassium 4.8 mEq/l and chlorine 108 mEq/l. Total pro-

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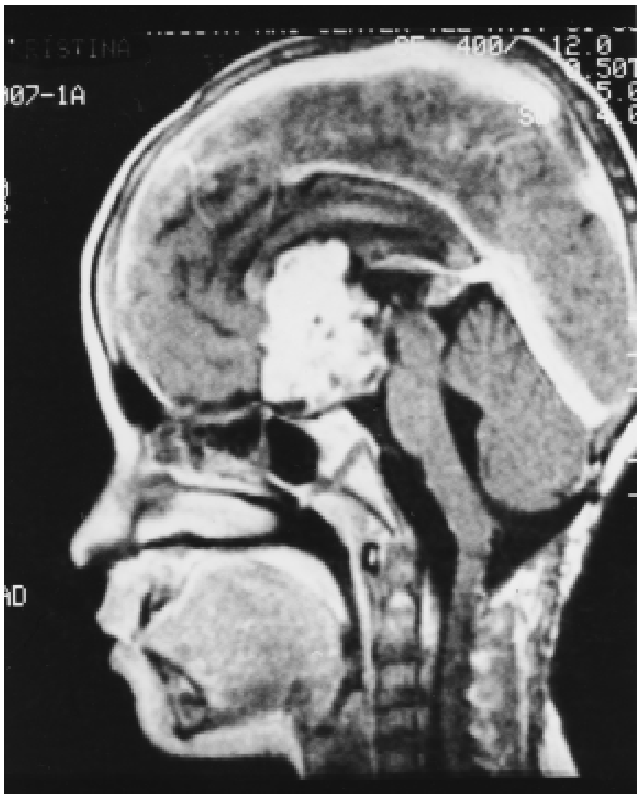

**Fig. 1.** The large chiasmatic/hypothalamic tumor.

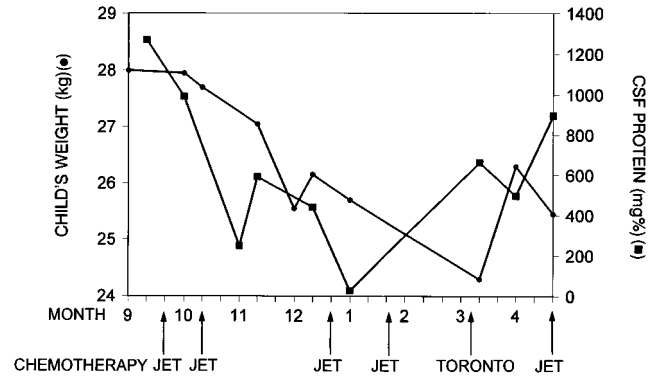


**Fig. 2.** The correlation between CSF protein and amount of ascitic fluid presented as body weight.

tein level was 7.4 g/dl, with albumin 4.7 g/dl. CSF protein level was 1,270 mg/dl with normal glucose and chlorine. Ascitic fluid was drained and showed a protein concentration of 1,800 mg/dl. There was no evidence of infection or tumoral spread in the peritoneal fluid. Due to the combination of high protein in both the CSF and peritoneal fluid, chemotherapeutic treatment with carboplatin, 1 gm/$m^2$ and VP-16 300mg/$m^2$, (JET regimen [4]) was resumed, on the assumption that following chemotherapy a reduction in secreted protein would lead to an improvement in the ascites. Later, treatment was changed to VP-16, 100 mg/$m^2$ × 2 days, ifosfamide 3 gm/$m^2$ × 2 days, and cisplatin 100 g/$m^2$. This is a modification of treatment protocol developed in Toronto with cisplatin used instead of carboplatin (Toronto Protocol). With chemotherapy, the extent of the ascites, as measured by weight and abdominal girth, dropped, in good correlation with the CSF protein level (Fig. 2). During this period, marked elevations of serum sodium were noted, fluctuating from 153 to 190 mEq/l. The girl had very little desire to drink, even during periods of high serum sodium concentrations, with only a small fluid intake. No abnormalities on kidney function tests were observed. Urinalysis was normal. Despite high serum sodium concentrations, urine osmolarity remained around 270 mOsmol/kg. No neurological symptoms or signs were noted, and the girl remained alert and coherent even with very high sodium levels; on the contrary, two separate episodes of a marked decrease in level of consciousness were associated with sodium levels of 131 and 137 mEq/l. Two years after initial presentation, the girl died from fungal pneumonia during postchemotherapy leukopenia.

## DISCUSSION

Ascites is a known complication of ventriculoperitoneal shunting; about 20 cases have been previously reported [1,5–7]. In such cases, an infection or tumoral spread via the shunt tubing should be ruled out [8,9]. High CSF and ascitic fluid protein were found by Adegbite and Khan [6] in an 11-year-old with craniopharyngioma. La Ferla et al. [5] described a 26-year-old woman with right temporal fibrillary astrocytoma, shunt and protein-rich ascites [5]. However, most of the reported cases had an optic pathway tumor. Yount et al. [7] described two children with optic nerve glioma and ascites in whom the protein content of the ascitic fluid was high, but that of the CSF was not. Kretschmar et al. [3] presented another infant with extensive optic pathway tumor and ascites, which responded to chemotherapy and resolved [2]. West et al. [10] described three children with optic pathway tumor associated with ascites following ventriculoperitoneal shunt placement. They suggested that because the tumor is widely exposed to the CSF space, protein exuded by the mass into the subarachnoid space would cause an elevated CSF protein concentration. The present child also had high protein ascitic fluid. An elevation in the protein concentration of the CSF may result from a defective blood-brain barrier in the tumor blood vessels or from protein production by the tumor itself. Astrocytomas are capable of producing specific proteins, such as astroprotein [1,11]. The high protein can contribute to the formation of ascites via both interference of absorption by the arachnoid villi and the peritoneum, and osmotic trapping of fluid in the peritoneal cavity [1]. La Ferla et al. [5] suggested that the formation

of a sclerosing peritonitis was the cause of the nonabsorption of the fluid. The high protein content of the CSF responded to chemotherapy, and so long as the tumor responded to the chemotherapy, the ascites improved. Similar responses were also noted by Kretschmar and Linggood [3] in their patients with OCG. While some authors [10] recommended that in such cases the shunt should be converted to a ventriculoatrial system, it may also be suggested that a further trial to reduce the tumor mass with chemotherapy may be done before applying neurosurgical techniques such as replacement of the shunt to the atrium or gallbladder.

Of interest is the marked hypernatremia with the absence of any of the neurological signs and symptoms which usually accompany high sodium concentrations. Patients with similar degrees of hypernatremia may be expected to present with circulatory failure, muscular asthenia, disorientation, convulsions, coma and other cerebral symptoms [12]. However, in the present case, even with high sodium concentrations, the child behaved normally; in fact, decreases in her level of consciousness were associated with normal serum sodium. There was no polyuria or polydipsia, and the clinical picture was compatible with hypodipsic hypernatremia. Several patients with suprasellar tumors have been reported with similar clinical picture. It is suggested that damage to the hypothalamic osmoregulatory mechanism was the cause of the hypodipsia and the resultant hypernatremia [13,14].

Tang et al. [1] described an infant with chiasmatic glioma, inappropriate secretion of antidiuretic hormone (IADH), nonobstructive hydrocephalus and chronic ascites following ventriculoperitoneal shunting. They related both the hydrocephalus and the ascites to the high protein content of the CSF. In contrast to our patient, theirs had hyponatremia.

The association between optic pathway glioma, hydrocephalus and ascites seem to be more than coincidental. In some cases, as in ours, the ascites responded to chemotherapy and a shunt replacement could be avoided. The association between the electrolyte abnormalities and the high protein CSF remains speculative. It may be that the high CSF protein alters the hypothalamic osmoreceptor function, thus causing hypernatremia. Furthermore, the question of whether it is the high protein content per se or a specific protein that alters CSF absorption is still unanswered. Presumably, physicians' awareness of this association and the work-up of more such cases will improve our understanding of the mechanisms involved.

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